VIRGINIA DEPARTMENT OF HEALTH
Protecting You and Your Environment

Protocol for Primary Healthcare Providers
Virginia Early Hearing Detection and Intervention Program
Virginia Department of Health

This document provides guidance and recommended procedures for primary healthcare providers (PHP) to implement requirements that are specified in the *Code of Virginia*, Section 32.1-46 [1] and *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System*[2].

These protocols represent the best practice that the Virginia Early Hearing Detection and Intervention Program (VEHDIP) Advisory Committee recommends based on the policy statement *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing* and other relevant sources such as the Centers for Disease Control and Prevention (CDC), and National Center on Birth Defects and Developmental Disabilities. The VEHDIP Advisory Committee consists of representatives from relevant groups including, but not limited to, primary care physicians, otolaryngologists, audiologists, speech pathologists, nurses, and parents.

It is important to recognize that newborn hearing screening is only one component of a comprehensive approach to the management of childhood hearing loss. The process also requires follow-up diagnostic services, counseling, intervention programs, and parental education. This comprehensive process should be administered by a multidisciplinary team including, but not limited to, audiologists, physicians, educators, speech/language pathologists, nurses, and parents.

VEHDIP goals are to identify congenital hearing loss by 3 months of age following the CDC *1-3-6 methodology*:

1 – All newborns will be screened for hearing loss **before 1 month** of age.

3 – All newborns who have failed their hearing screen will receive a diagnostic evaluation **before 3 months** of age.

6 – All infants diagnosed with hearing loss will be enrolled in early intervention services **before 6 months** of age.

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[1] To access the *Code of Virginia* citation, go to: http://leg1.state.va.us/cgi-bin/legp504.exe?000+cod+32.1-64.1
[2] To access the *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System,* go to http://leg1.state.va.us/000/reg/TOC12005.HTM#C0080

I. Virginia Early Hearing Detection and Intervention Program Overview

Providers performing initial hearing screening (typically hospitals) are required to perform this screen prior to discharge, and report results to the Virginia Department of Health (VDH) and to the primary healthcare provider from whom the infant will receive care after discharge. They must also provide written information to the parent that includes purposes and benefits of newborn hearing screening, the procedures used for screening, recommendations for further testing, and where testing can be obtained.

Best practice guidelines for providers performing initial screening include:

- Report screening results to the VDH, and to the primary healthcare provider, within two weeks of discharge.
- Assess all infants for risk indicators associated with hearing loss.

Persons providing audiological services to infants after hospital discharge are required to provide the screening or evaluation results to the parent or guardian and to the child's primary healthcare provider. They must also give resource information to the parent (or guardian) of any child found to have a hearing loss, including but not limited to, the degrees and effects of hearing loss, communication options, amplification options, the importance of medical follow-up, and agencies and organizations that provide services to children with hearing loss and their families. Finally, they must send the Audiological Reporting Form—including test results, diagnosis, and recommendations—to the VDH within two weeks of the patient visit. The Audiological Reporting Form, including risk indicators and instructions, can be downloaded from the VDH website www.vahealth.org/hearing.

Best practice guidelines for person providing audiological services to infants after hospital discharge include:

- Provide the hearing screening or evaluation results to the parent or guardian at the time of the visit.
- Assess infants and children for risk indicators associated with hearing loss.
- Provide the hearing screening or evaluation results to the primary healthcare provider within two weeks of the patient visit.
- Refer the family to Part C Early Intervention and/or other educational programs.

The VDH Virginia Early Hearing Detection and Intervention Program is required to collect, maintain, and evaluate hearing screening data. Program staff must provide follow-up, including communicating with the parent or guardian to assure that they have the information needed to seek timely and appropriate follow-up services. They must also provide training and technical assistance to hospitals, and communicate critical performance data to hospitals yearly. Lastly, the program is required to review and evaluate the surveillance system including follow-up rates, false-positive rates, false-negative rates, referral mechanisms and effectiveness of tracking.

Best practice guidelines for the VEHDIP include:

- Monitor hospital data and rate of reporting.

- Provide epidemiological analysis of the data for planning and program management purposes.
- Approve centers/providers of diagnostic audiological services for infants and young children under this program.
- Provide training and technical assistance to birthing centers.

Primary healthcare providers should:

- Receive and review the hearing screen results, risk indicator findings, and diagnostic evaluation results from hospitals, audiological providers, and VDH.
- Refer infants to an audiologist for follow-up or diagnostic assessment, as needed.
- Receive and review information from VDH regarding available resources to assist practitioners and families with a child at risk for, or diagnosed with, hearing loss.
- Provide medical assessment as recommended in this guidance document.
- Provide regular surveillance of developmental milestones, auditory skills, parent or guardian concerns, and middle-ear status consistent with the American Academy of Pediatrics (AAP) recommended periodicity schedule. All infants and toddlers should have an objective developmental screen with a standardized tool at 9-, 18-, and 24-to-30 months of age.
- Refer infants and toddlers who do not pass the speech-language portion of a standardized developmental screen, or whose behavior/responses cause concern regarding hearing or language, for speech-language evaluation and audiology assessment.
- Refer infants diagnosed with hearing loss to Part-C Early Intervention services. Visit the Infant & Toddler Connection of Virginia website www.infantva.org for information on referral processes and contacts.

II. Interpretation of Hearing Screen Results

Table I summarizes information on hearing screen results and associated primary healthcare provider (PHP) actions.

Table I: Interpretation of Hearing Screen Results

Screening Result	Interpretation by PHP	PHP Follow-up Recommendations
Passed Screen	Infant passed in both ears	Monitor communication and language development.
Passed with Risk	Infant passed in both ears but is at risk for progressive or late onset hearing loss	Inform the parent of the need for a diagnostic audiological assessement by 24 months of age and refer as needed. Perform developmental surveillance and screening consistent with American Academy of Pediatrics recommendations.
Missed Screen	Infant did not have a required hearing screen prior to discharge	Reinforce with parent the need to assure screening is done. The hospital is required to make arrangements for the infant to be screened as soon as possible but no later than 1 month of age. (Hospitals are responsible for providing mechanism by which screening can occur at no additional cost to the family).
Failed Screen	Infant did not pass in one or both ears	Infant should be tested again within a month of hospital discharge. Reinforce this need with the parent. Follow-up should be done at a VDH-approved facility for diagnostic audiological assessment (a list is available at **www.vahealth.org/hearing**). Note: Some hospitals have the family return to the hospital for a re-screen; if the infant fails again, refer to an audiologist for an assessment before 3 months of age.
Infant not born in a hospital	Any infant born outside of hospital facility, e.g., home birth	Refer for initial screening as soon as possible after birth to a VDH-approved facility for diagnostic audiological assessment.

A flow chart depicting the newborn hearing screening and follow-up process is in **Attachment 1**.

III. PHP Recommended Actions for Infants With Confirmed Hearing Loss

Every infant with confirmed hearing loss needs medical evaluation to determine the etiology of hearing loss, identify related physical conditions, and make recommendations for treatment and referrals to other resources. The primary healthcare provider should thoroughly document family history and the mother's prenatal risk factors. This will support ongoing care for the affected child as well as monitoring other children in the family. Document:

- Prenatal conditions: ototoxic medication exposure, pregnancy complications, immunization status of mother for rubella, maternal status for syphilis, maternal drug and/or alcohol use, and history of frequent spontaneous abortions.
- Family history: hereditary childhood sensorineural hearing loss and family members with permanent hearing loss with onset before age 30 not related to trauma or medical condition.

Complete a physical examination with special attention to:

- Minor anomalies: unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance.
- Major anomalies: dysmorphic features that cause significant cosmetic or functional abnormality, such as cleft palate, cardiac, limb, or other skeletal deformities.
- Poor growth, microcephaly, or abnormal neurological exam.

Obtain laboratory and imaging studies:

- Urine culture for cytomegalic inclusion virus before aged 3 weeks if possible.
- Consistent with history of findings, consider testing for rubella, syphilis, or toxoplasmosis.
- Chromosomal studies if significant dysmorphic features.
- EKG if cardiac condition suspected.
- Skeletal survey if growth delayed or disproportionate.
- Head CT or MRI if neurological exam abnormal.

Refer as follows:

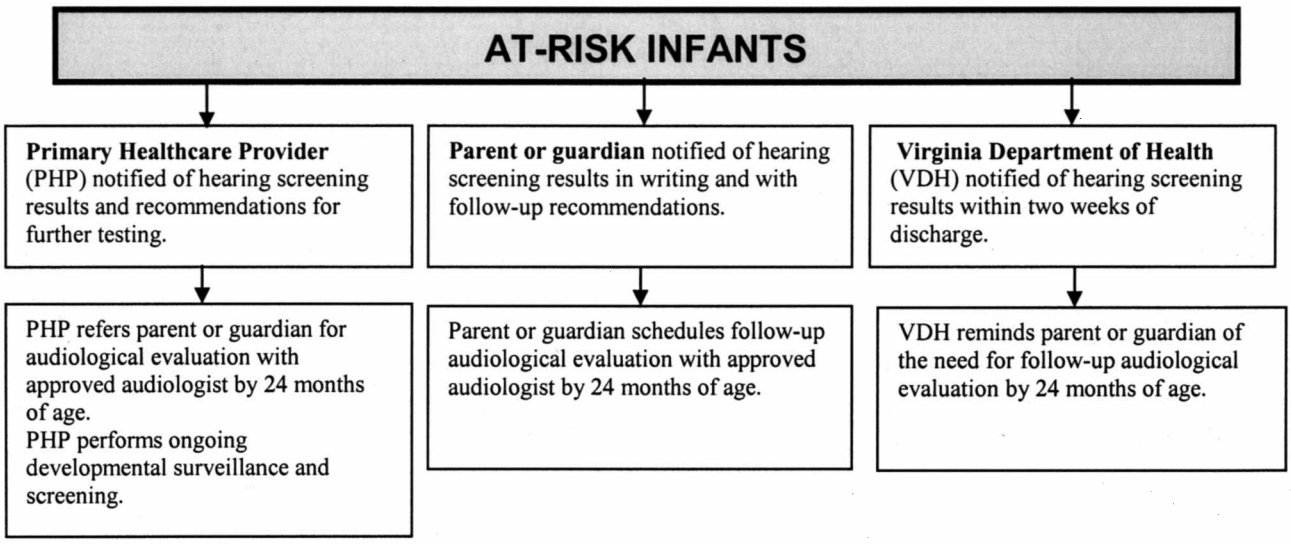
- To otolaryngology (ENT).
- To ophthalmology/cardiology/nephrology evaluation if indicated.
- For genetic evaluation and counseling for both syndromic and non-syndromic forms of hearing loss.
- Siblings with increased risk of having hearing loss for an audiological evaluation.

IV. Risk Indicators

Birth hospitals in Virginia are responsible for determining the risk status for hearing loss on every newborn regardless of the results of the hearing screen. Risk-status data assist with monitoring for progressive, delayed-onset, and/or conductive hearing loss. VDH recognizes the risk indicators identified by the policy statement *Year 2007 Position Statement: Principles and Guidelines for Early Detection and Intervention Programs, Joint Committee on Infant Hearing* (See **Attachment 2**).

Some indicators may not be determined during the course of the hospital stay. Therefore, infants and young children who have late-onset or late-identified risk indicators should be monitored for speech, language, and hearing developmental milestones by the primary healthcare provider during well-child visits.

The following diagram summarizes processes that should be followed to ensure infants and children who are at risk for hearing loss receive appropriate audiological follow-up.



Some of these indicators are not present and/or would not be identified in the newborn period. These include parental concern and some neurodegenerative disorders or sensory motor neuropathies. These are included in the risk indicator list because parents and physicians should be informed about all indicators that can contribute to development of hearing loss beyond the newborn period.

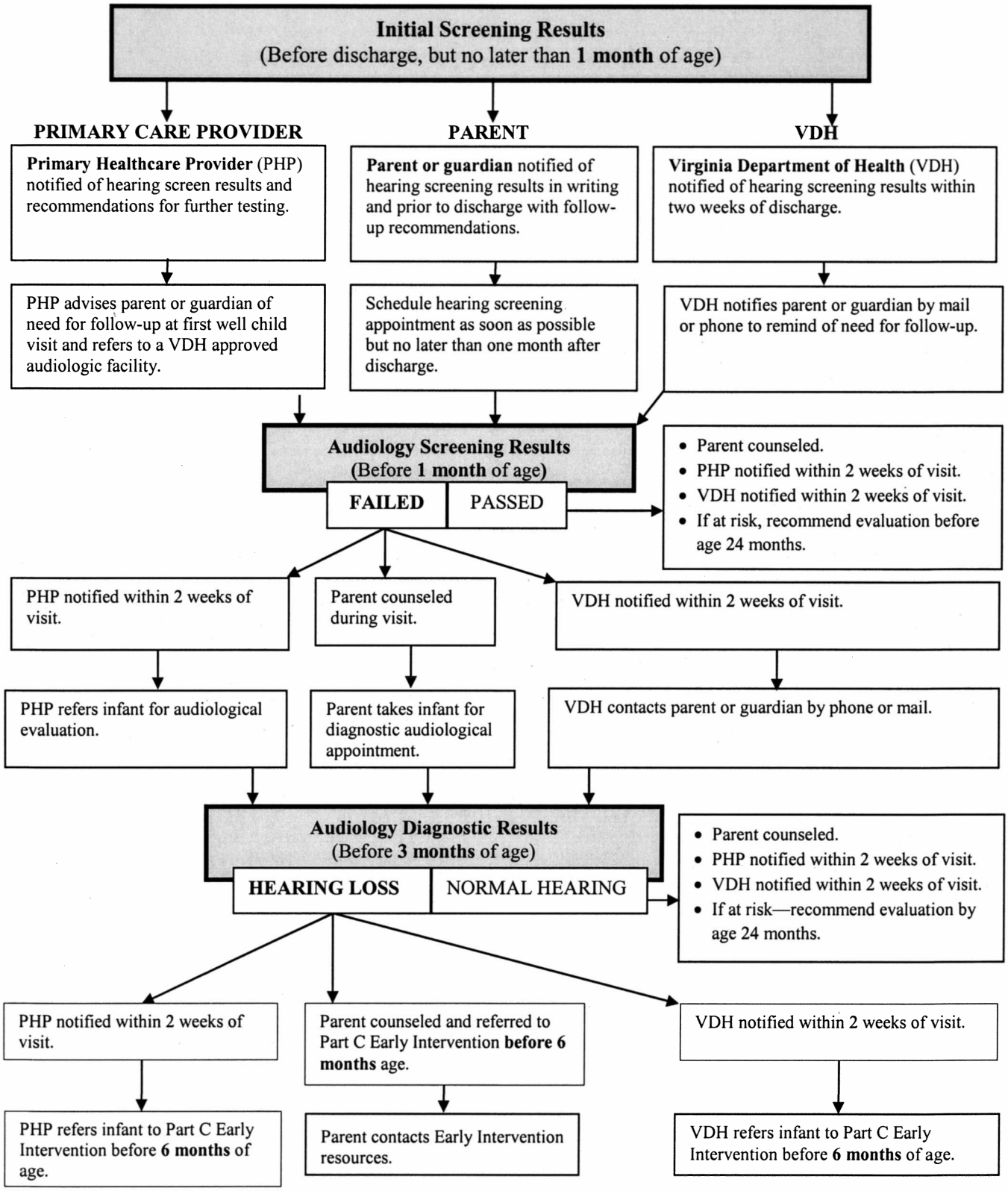
Infants who pass the hearing screen but have an identified risk indicator for progressive or delayed-onset hearing loss (**pass with risk**) should have a complete diagnostic evaluation by 24 months of age.

V. Contacts

For more information or further assistance, families are encouraged to contact:
Virginia Department of Health
Office of Family Health Services
Virginia Early Hearing Detection and Intervention Program
109 Governor Street, 8th Floor
Richmond, VA 23219

Phone: Toll Free 1-866-493-1090 TTY 7-1-1
Fax: 804-864-7721
Website: **www.vahealth.org/hearing**

VI. Attachment I: Virginia Early Hearing Detection and Intervention Program Process Flow Chart



Attachment 2: Risk Indicators for Progressive or Delayed-Onset Hearing Loss

(For Use with Neonates and Infants Through 2 Years of Age)

Family history of permanent childhood hearing loss

- Mother of child
- Father of child
- Sister of child
- Brother of child
- Grandmother of child
- Grandfather of child
- Aunt of child
- Uncle of child
- 1st cousin of child
- More than one relative of the same parent

Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction

- Branchio-oto-renal (BOR)
- Noonan
- CHARGE association
- Pierre Robin
- Rubenstein-Taybi
- Stickler
- Williams
- Zellweger
- Goldenhar (oculo-auriculo-vertebral or OAV)
- Trisomy 8 – Warkany syndrome
- Trisomy 21 – Down syndrome
- Trisomy 18 – Edwards syndrome
- Trisomy 13 – Patau syndrome
- Trisomy 9 – Mosaic syndrome

Postnatal infections associated with sensorineural hearing loss

- Confirmed bacterial meningitis
- Confirmed viral meningitis

In utero infections

- Cytomegalovirus
- Herpes
- Rubella
- Syphilis
- Toxoplasmosis

Neonatal indicators

- Intensive care greater than (>) 5 days
- Extracorporeal membrane oxygenation (ECMO)
- Exposure to ototoxic medications: at risk aminoglycoside exposure
- Mechanical ventilation
- Hyperbilirubinemia requiring exchange transfusion

Syndromes associated with progressive hearing loss

- Neurofibromatosis
- Osteopetrosis
- Alport
- Jervell & Lange-Nielson
- Waardenburg
- Pendred
- Usher

Neurodegenerative disorders, such as

- Hunter syndrome
- Charcot-Marie-Tooth syndrome
- Friedreich's ataxia

Head trauma requiring hospitalization

- Basal skull/temporal bone fracture

Other – specify if chosen

Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay

Craniofacial Anomalies

- Pinna
- Cleft palate
- Atresia
- Microtia
- Choanal atresia
- Temporal bone anomalies

Chemotherapy

Based on *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing.*

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Approved by:

Karen Remley, MD, MBA, FAAP
State Health Commissioner

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Date